

Case report: cobblestone

Superimposed primary chancre in a patient with Adamantiades-Behçet's disease

N Meljanac, E Dippel, Ch C Zouboulis

Adamantiades-Behçet's disease was diagnosed in a 42 year old Turkish patient with recurrent oral aphthae, genital ulcerations, papules, and sterile pustules, histologically presenting as cutaneous vasculitis, and intermittent arthritis with joint effusion particularly of the knees. Six months after initial improvement under treatment with colchicine 2 mg/day, a solitary genital ulcer with enlarged inguinal lymph nodes appeared and persisted for 7 weeks despite the continuation of colchicine treatment and the introduction of clindamycin 2 mg/day intravenously. The unusual persistence of the ulcer and the failure of clindamycin therapy led to further differential diagnostic considerations and the identification of primary syphilis. The genital lesion healed 4 weeks after initiation of treatment with tetracycline 2 mg/day by mouth for 15 days.

(Sex Transm Inf 1999;75:124–125)

Keywords: Behçet's disease; syphilis

Case report

A 42 year old Turkish man presented on May 1997 at the department of dermatology at the Free University of Berlin with several 2–10 mm large buccal aphthae, a 2 mm large ulcer at the orificium urethrae, a 1 cm wide ulcer on the penile shaft (fig 1a), and papulopustular skin lesions at his shoulders and shanks resembling folliculitis. There was a history of recurrent oral aphthae and genital ulcerations since October 1996 and recurrent erythematous papules and sterile pustules occurring particularly on the extremities since December 1996. Recurrent conjunctivitis and intermittent mono- or polyarthritis with joint effusion, especially of the knees, completed the clinical picture. Family history was negative.

The combination of clinical findings led to the initial diagnosis of Adamantiades-Behçet's disease. Ophthalmological examination was normal. A pathergy test was negative. HLA type I analysis revealed the alleles A2, A24,

B51, B18, Bw4, Bw6, and Cw7. Further laboratory values were as follows: cardiolipin autoantibodies of the IgG type 11.9 anticardiolipin antibodies IgG units/ml (GPL-U/ml) (normal values <12 GPL-U/ml), of the IgM type 3.2 anticardiolipin antibodies IgM units/ml (MPL-U/ml) (normal <6 MPL-U/ml), antinuclear antibody titre 1:80, anti-herpes simplex virus titre 1:40 in the complement fixation test with positive IgG and negative IgM antibodies in enzyme immunoassay. Herpes simplex virus polymerase chain reaction (PCR) and cytomegalovirus PCR, *Chlamydia trachomatis* ELISA, HIV antibodies, TPHA, and VDRL tests were negative. Hepatitis serology showed IgG antibodies to hepatitis A virus, but no indication of an active process. Unspecific inflammatory factors were elevated—namely, erythrocyte sedimentation rate 74 mm Hg in the first hour, C reactive protein 45 mg/l (normal <10 mg/l), and α_2 globulin in serum electrophoresis varying from 10.1 to 11.1% (normal 5–10%).

Skin biopsies from the genital ulcer and folliculitis-like lesions on the left shoulder and left shank were suggestive of perivascular inflammatory reaction and nodular vasculitis, findings compatible with Adamantiades-Behçet's disease. Systemic treatment with colchicine 1 mg twice daily by mouth was initiated which led to clearance of the mucocutaneous lesions within a period of 3 weeks.

In September 1997 the patient was again referred to our department with a 7 week history of a solitary genital ulcer. On examination, a 2 cm large ulcer of the penile shaft was detected, covered with a haemorrhagic crust, sharply demarcated, and surrounded by oedematous tissue. Enlarged, painless inguinal lymph nodes were also present (fig 1b). Oral mucosa and eyes were not affected. The patient reported an active sexual life with several female partners.

A bacterial superinfection of a genital lesion of Adamantiades-Behçet's disease was suspected and systemic treatment with clindamycin

Department of
Dermatology,
University Medical
Center Benjamin
Franklin, The Free
University of Berlin,
Berlin, Germany
N Meljanac
E Dippel
Ch C Zouboulis

Correspondence to:
Priv-Doz Dr Christos C
Zouboulis, Department of
Dermatology, University
Medical Center Benjamin
Franklin, The Free
University of Berlin,
Hindenburgdamm 30,
D-12200 Berlin, Germany.

Accepted for publication
18 January 1999



Figure 1 (a) A 1 cm large ulcer on the penile shaft of the patient at his first admission. (b) A 2 cm large ulcer of the penile shaft with asymptomatic enlarged lymph nodes (arrow) on the second referral.

3 × 600 mg intravenously was initiated. Colchicine treatment was continued. With this therapeutic regimen over 7 days the genital ulcer showed no tendency to heal. Another histopathological examination revealed an unspecific granulomatous reaction. With respect to the persistence of the lesion tests for syphilis were repeated with following findings: TPHA test 1:640, VDRL test 1:128, and FTA-ABS test positive. Because of a history of a penicillin hypersensitivity reaction the patient was treated with tetracycline 4 × 500 mg/day for 2 weeks. A complete healing of the ulcer was noted 4 weeks after initiation of treatment. The suspect female partner was identified, found positive for syphilis, and treated successfully.

Discussion

Our patient presented with clinical and histopathological features suggesting Adamantiades-Behçet's disease.¹ Since mucocutaneous lesions do not usually persist over 4 weeks, the 7 week old genital ulcer led to further diagnostic steps which detected the additional primary syphilis.

Gnad² and Sarica *et al*³ reported two patients with uveitis and uveitis combined with mucocutaneous lesions, respectively, as a result of secondary syphilis, who were misdiagnosed as having Adamantiades-Behçet's disease. In our case Adamantiades-Behçet's disease was diag-

nosed 6 months before the confirmation of primary syphilis, while syphilis serology was negative at that time.

This case emphasises the need for re-evaluating a chronic disorder with uncommon features of apparently typical lesions during its course in order to avoid misdiagnosis. It also demonstrates the importance of taking a sexual history of every genital lesion and the need to perform TPHA and VDRL tests with every atypical genital ulcer, irrespectively of earlier established diagnosis.⁴ Since Adamantiades-Behçet's disease is a chronic disorder with common monosymptomatic onset and recurrence,¹ there must be awareness of the possibility of additional diseases with similar symptomatology; dermatovenereological consideration of every genital lesion is especially important.

We thank Dr Peter K Kohl, Department of Dermatology and Venereology, Academic Hospital Neukölln, for critical review of the manuscript.

- 1 Zouboulis ChC. Adamantiades-Behçet's disease: clinical and experimental findings in 53 patients from the Berlin area. Habilitation thesis. Berlin: Medical Faculty of the Free University of Berlin, 1995.
- 2 Gnad HD. Zuweisungsdiagnose "Morbus Behçet". *Klin Monatsb Augenheilkd* 1975;175:548–50.
- 3 Sarica R, Azizlerli G, Közer LB, *et al*. Uveitis als Manifestation der Lues II-fehldiagnostiziert als Morbus Behçet. *Derm Monatsschrift* 1993;179:59–63.
- 4 Joyce JP, Waldman PC, Hood AF. Persistent genital ulceration. Chancroid. *Arch Dermatol* 1989;125:552–6.